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Case Report

Hodgkin's disease and bone marrow compression following a diagnostic delay in the Internal Medicine and Medical Oncology Unit at the National Teaching Hospital Center (NTHC)-Hubert Koutoukou Maga (HKM) of Cotonou: a case report and review of the literature

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Abstract

Introduction: Hodgkin's disease is a rare disease of the lymphoid tissue that occurs mainly in young adults. Its localization in the bone marrow is rare and can lead to severe functional disability. We are reporting in this work the case of a young boy of 18-year-old who was followed in the department. Observation: 18-year-old patient admitted to the department for exploration of febrile adenopathies (PDA), context of weight loss. The onset was reported to be one year prior to admission. The diagnosis of tuberculosis was suspected and treated for 4 months without success, in front of evocative signs with a notion of contagion. This situation was associated with moderate asthenia, intermittent fever without cough, or night sweats. The occurrence of a progressive paresis of the right pelvic limb, three days before admission, associated with low spinal pain and hyperesthesia of the pelvic limbs motivated her hospitalization. General conditions altered with a WHO performance index of 2. Temperature was 37°8C, HR 90bpm, Blod Pression (BP): 120/85mmHg, Respiratory Rate (RR) 20cpm, oxygen saturation (SpO_2): 98% on room air. Physical examination: presence of bilateral superficial cervical, axillary and inguinal PDAs, painless, mobile, of variable size. Paraplegia was noted with 3/5 motor strength in the left pelvic limb and 2/5 in the right pelvic limb. Low spinal pain of the burning type 9/10 according to VAS. Biology showed a major inflammatory syndrome. On imaging, the thoracoabdominopelvic CT scan showed hepatosplenomegaly and multiple deep retroperitoneal adenopathies with soft tissue infiltration. Spinal cord MRI revealed multiple spondylitis, an extensive intraductal and epidural tissue process at T12-L1 and L2-L3, and an anterior epidural infiltration opposite L2 responsible for spinal cord compression. Pathology of the cervical lymph node revealed the presence of Reed Stenberg cells with CD30+ expression on immunohistochemistry. The patient was put on chemotherapy and the evolution at 3 months was slowly favorable with progressive functional recovery. Conclusion: Spinal cord compression is a rare complication of Hodgkin lymphoma. It often occurs at an advanced stage of the disease. The evolution can be favorable if treated early and appropriately.

Keywords: Lymphoma, Hodgkin, diagnostic delay, bone marrow compression.

INTRODUCTION

Hodgkin's disease, also known as Hodgkin's lymphoma or malignant lymphogranulomatosis (Yung et al., 2003), is a rare malignant disease of the reticuloendothelial tissue characterized by the presence of a particular cell called Reed Sternberg cell. It occurs mainly in young adults, adolescents, and older children and has two peaks in frequency, between 20 and 30 years of age and over 50 years of age (Georg et al.,1983 ; d'Andon et al., 2003. Spinal cord compression is a rare complication of Hodgkin's disease, usually occurring in 5% of cases, in the context of progressive and advanced disease (Susan et al., 1995). Hodgkin's disease can be revealed exceptionally by spinal cord compression, causing neurological signs without other systemic involvement (Lyding et al. 1987, Rabhi et al. 2007). The diagnosis of this location may be difficult in the presence of nonspecific neurological symptoms, radiographs, and normal CSF analysis. Thus, many patients may develop complications before the diagnosis is made with poor survival despite the effectiveness of available treatments. Although spinal cord compression is rarely described in the literature as a complication of Hodgkin's disease
(Rabhi et al. 2007), in the context of limited resources, diagnostic errors can lead to delayed management and consequently to serious and irreversible neurological complications.
We describe here the case of a young patient with spinal cord compression in whom the diagnosis of Hodgkin's disease was made and confirmed in the internal medicine and medical oncology department of the CNHU-HKM of Cotonou.

Observation

This was an 18-year-old man, with no known pathological history and any notion of documented familial cancer nor exposure to a particular risk, admitted to the hospital for exploration of a febrile adenopathy. The onset was one year prior to admission with moderate asthenia and involuntary weight loss of about 4kg in 10 months. The diagnosis of tuberculosis was suspected and treated for 4 months without success, in front of evocative signs with a notion of contage, without bacteriological confirmation. The treatment was then stopped due to the absence of clinical improvement with the appearance of other superficial adenopathies, especially axillary and inguinal, and intermittent fever without cough or nocturnal hypersudation. Three days before admission to the department, the patient presented with a slow onset of right pelvic limb paresis with a sensation of paresthesia.
The physical examination revealed a temperature of 37.8°C, bilateral superficial cervical, axillary and inguinal adenopathies of firm consistency, painless, mobile in relation to the deep and superficial plane, the most voluminous in the right latero-cervical region measuring 5 cm in the long axis (Figure 1)
On neurological examination, we noted a paraplegia with muscle strength rated at 3/5 on the left and 2/5 on the right, a decrease in osteotendinous reflexes in the pelvic limbs, and diffuse burning pain in both legs rated at 9/10 on the numerical scale. There was no hepatomegaly or splenomegaly. The cardiovascular, ENT, and genitourinary examinations were unremarkable.
On the paraclinical level
The blood count showed microcytic anemia with a hemoglobin level of 8.4 g/L, mean corpuscular volume (MCV) of 70.4 fl, mean corpuscular hemoglobin concentration (MCHC) of 30%, mean corpuscular hemoglobin content (MCHC) of 21.3 pg. A predominantly neutrophilic hyperleukocytosis with leukocytes at 24.8G/L with neutrophils at 24g/L, lymphocytes at 0.49 g/L, monocytes at 0.24 g/L and thrombocytosis at 599 g/L.
CRP was highly elevated at 392mg/L and the sedimentation rate accelerated to 56 mm in the first hour. Serum protein electrophoresis in favor of an inflammatory reaction without monoclonal peak. LDH was 329IU/L. There is no hepatic cytolysis. Aspartate-Aminotransferase (ASAT) 15IU/L, Alanine Aminotransferases (ALAT) 21IU/L, Gamma GT (GGT) 50IU/L, Alkaline Phosphatase 63IU/L. Total bilirubin was normal 6 mg/L, conjugated bilirubin 3 mg/L.
HIV serology was negative, as well as VDRL and TPHA, hepatitis viral serologies HBV and HCV were also negative. The search for *Mycobacterium tuberculosis* DNA by GenXpert on gastric tubing fluid was also negative.
On imaging, the Thoracic-Abdominal-Pelvic (TAP) scan showed homogeneous hepatosplenomegaly (Figure 2), multiple retroperitoneal adenopathies and soft tissue infiltration (Figure 3).
Spinal cord MRI revealed multiple spondylites of the C2, C3 vertebrae (Figure 4). L1, L2, L5, the sacrum (Figure 5). There were necrotic areas on L2 and S1 vertebrae, extensive epidural intraductal tissue process of T12-L and L4-L5.
In addition to these lesions, there was an anterior epidural infiltration opposite L2 responsible for compression of the cone and dural sheath. (Figure 5).
Pathological examination of the cervical lymph node biopsy showed a lymphoproliferation of the nodular architecture, with nodules separated by fibrous septa (figure 6). These tumor cells correspond to Reed Stenberg cells and lacunar cells, the background is made of small lymphocytes (figure 7).
On immunohistochemistry, the tumor cells expressed CD30. No expression of CD15 and ALK (Figure 7). This is in favor of a classical sclerodular Hodgkin's lymphoma. Thus, the diagnosis of Ann Arbor stage IIIEB Hodgkin's lymphoma was made.

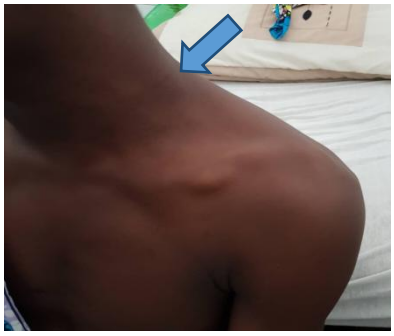

Figure 1. Right laterocervical adenopathy.

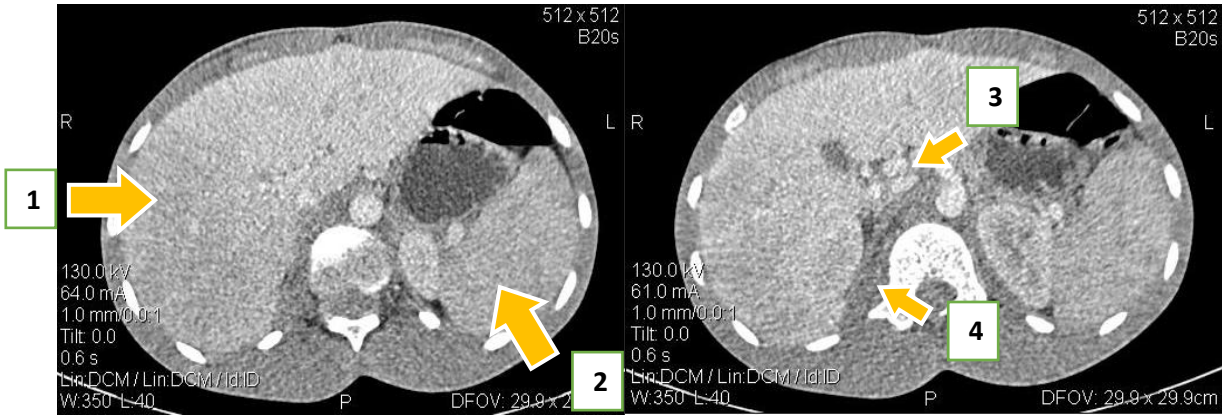

Figure 2
1- Hepatomegaly
2- Splenomegaly

Source : Internal Medecine - Medical Oncology CNHU-HKM,Cotonou, Benin

Figure 3
3- Retroperitoneal adenopathy
4- Soft tissue infiltration

Source : Internal Medecine - Medical Oncology CNHU-HKM,Cotonou, Benin

The treatment started was chemotherapy with the ABVD (Doxorubicin-Vinblastine-Bleomycin-Dacarbazine) protocol. The patient also received a corticosteroid bolus of 120 mg per day for 3 days. Symptomatic treatment of pain was done with Morphine syrup and pregabalin. A thoraco-abdominal corset was made and gentle physical therapy was started to maintain limb mobility after 2 cycles of chemotherapy. (figure 8 a, b)
The evolution after these 2 cycles of chemotherapy is marked by a clinical improvement with apyrexia, regression of adenopathies, and progressive recovery of muscle strength, almost complete on the left and 3/5 on the right figure 8 (a, b).

DISCUSSION

The responsibility of Hodgkin's lymphoma in spinal cord compression is estimated at 5% and concerns advanced disease with diffuse lymph node and visceral extension (4 Susan et al., 1995). Paul et al., (2003) described 12 cases

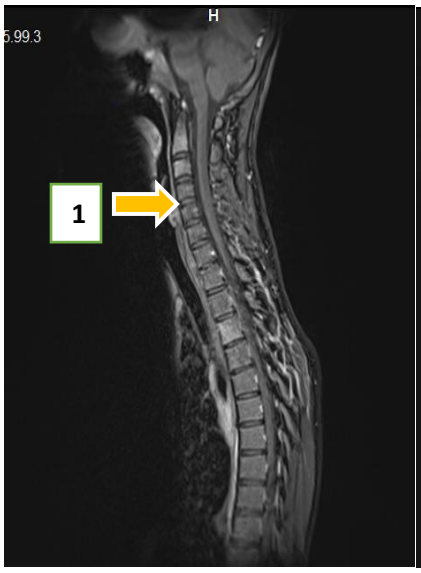

Figure 4

- 1 Spondylitis C2, C3

Source: InternalMedicineMedicalOncology CNHU-HKM, Cotonou, Benin

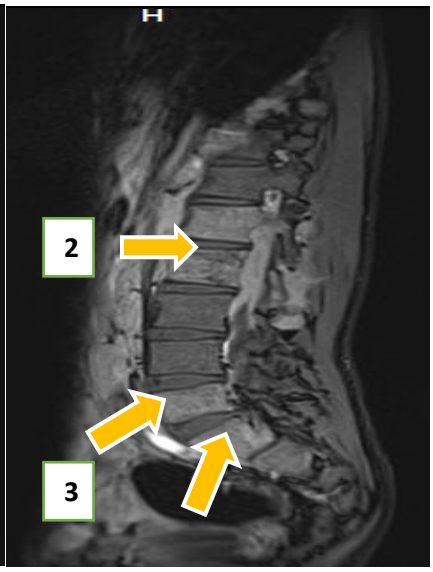

Figure 5

- 2 Spondylitis T12
- 3 Spondylitis L 2, L3, L4

Source: InternalMedicineMedicalOncology CNHU-HKM, Cotonou, Benin

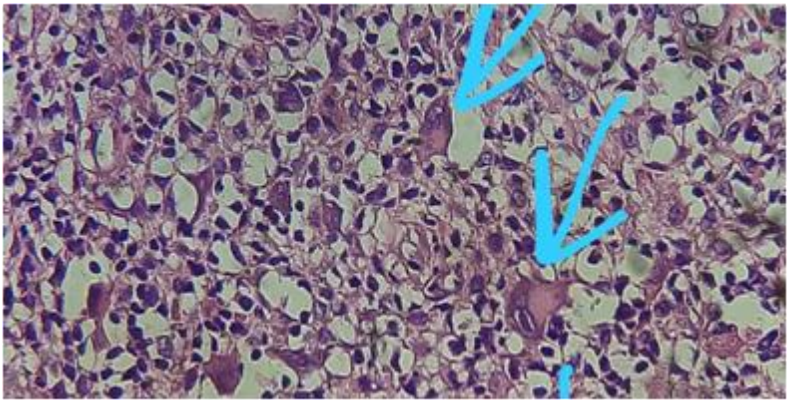

Figure 6

- Presence of Reed Sternberg cells

Source: Dubois anatomopathologylaboratory Cotonou, Benin

of spinal cord compression revealing Hodgkin's lymphoma, among which 8 presented adenopathy with other visceral localizations in addition to spinal involvement (Paul et al., 2003).

Spinal cord involvement in Hodgkin lymphoma may originate either from hematogenous dissemination via arterial or venous plexuses or from contiguous development from retroperitoneal or thoracic lymph nodes, often resulting in Hodgkin epiduritis (Mireau et al. 2009). Infiltration and dissemination can also occur through the cerebrospinal fluid (CSF) along the Virchow-Robin spaces (perivascular space between the subarachnoid space and the neural parenchyma) (Jardin et al., 1999). In our patient, the mechanism is probably mixed with hematogenous dissemination and by contiguity of the retroperitoneal lymph nodes. The thoracic spine is the most involved

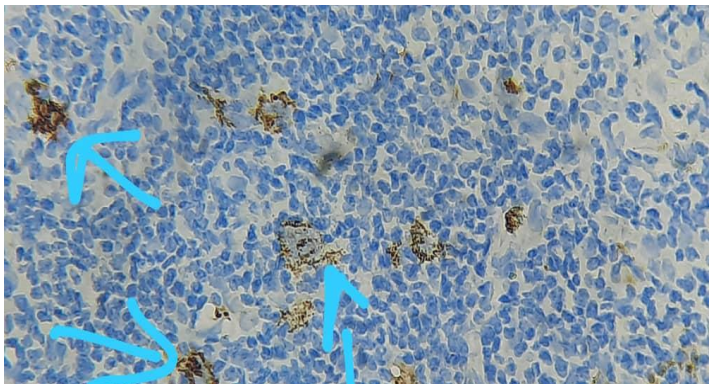

Figure 7

- Presence of brownish colored cells, expressing CD30

Source: Dubois Anatomopathology Laboratory Cotonou, Benin

Figure 8 a:patient seenfrom the front with the plastered corset

Figure 8 b:patient seenfrom the back with the plastered corset

region, followed by the lumbar region and then the cervical spine(Rabbi et al., 2007).Most cases of Hodgkin's lymphoma occur in people who do not have identifiable risk factors as in the case of our patient (Leukemia & Lymphoma Society, 2013).and the spinal cord involvement usually occurs in the advanced stage of the disease(Susan et al., 1995).

Regarding the clinical expression of Hodgkin's disease, the patient usually presents with painless cervical adenopathy that may be accompanied by weight loss, prolonged fever, and night sweats (Kasper et al., 2012; Uehara et al., 2013). These manifestations were present in this patient except for the night sweats.

Spinal cord involvement is a rare complication that is usually encountered in cases of delayed diagnosis of diffuse Hodgkin's disease or sometime after the initial diagnosis. Subsequently, back pain and neurological deficits occur depending on the level and location of the lesion [6 (Rabbi et al., 2007; Uehara et al., 2013). This symptomatology may mimic other conditions such as tuberculosis in our context, which may delay the diagnosis and initiation of appropriate treatment (Fourati et al., 2017). Our patient presented with similar signs after having been treated a year earlier as having tuberculosis without bacteriological confirmation, and without success.

Regarding imaging, MRI is a reference examination for

diagnosis, analysis of the extent, number of lesions, and impact on the caliber of the spinal canal and the spinal cord. The tumor tissue is most often hyposignal T1 and hypersignal T2. The tumor material is contrast-enhancing (often moderately) on injection (Mireau et al., 2009). A full-body CT scan is useful to complete the evaluation of disease extension (Ghedira et al. 2019).

For the treatment of Hodgkin lymphoma, chemotherapy is the gold standard and may be effective only if it is given early (Rabhi et al., 2007). However, surgical reduction is indicated in case of rapid worsening of symptoms or in cases of primary disease with spinal cord compression requiring histopathological diagnosis (Rabhi et al., 2007 ; Ghedira et al. 2019). Radiotherapy is useful for additional local control of epidural and paraspinal spread (Ghedira et al. 2019).

The evolution of Hodgkin lymphoma with spinal localization is globally favorable if the treatment is well conducted. Patients show functional recovery in 86% of cases (Rabhi et al., 2007) and complete clinical improvement in 61% of cases (Susan et al., 1995).

CONCLUSION

Spinal cord compression is a rare complication of Hodgkin lymphoma. It often occurs at an advanced stage of the disease and can lead to severe neurological consequences if not diagnosed early. The evolution is globally favorable if the treatment is well conducted, with a functional recovery in 86% of cases and a complete clinical improvement in 61% of cases.

Conflicts of interest: None

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